CORTEX 52 (2014) 75–85

Available online at www.sciencedirect.com

ScienceDirect

Journal homepage: www.elsevier.com/locate/cortex

Research report

When a loved one feels unfamiliar: A case study on the neural basis of Capgras delusion

Christiane M. Thiel [a,b,*], Sara Studte [a,c], Helmut Hildebrandt [a,d], Rene Huster [e] and Riklef Weerda [a]

[a] Biological Psychology Lab, Department of Psychology, European Medical School, Carl von Ossietzky Universität, Oldenburg, Germany
[b] Research Center Neurosensory Science, Carl von Ossietzky Universität, Oldenburg, Germany
[c] Experimental Neuropsychology Unit, Department of Psychology, Saarland University, Saarbrücken, Germany
[d] Department of Neurology, Hospital Bremen-Ost, Germany
[e] Experimental Psychology Lab, Department of Psychology, European Medical School, Carl von Ossietzky Universität, Oldenburg, Germany

ARTICLE INFO

Article history:
Received 19 June 2013
Reviewed 26 July 2013
Revised 21 October 2013
Accepted 2 December 2013
Action editor Stefan Schweinberger
Published online 12 December 2013

Keywords:
Capgras syndrome
Social cognition
Faces
Neuroimaging
fMRI

ABSTRACT

Perception of familiar faces depends on a core system analysing visual appearance and an extended system dealing with inference of mental states and emotional responses. Damage to the core system impairs face perception as seen in prosopagnosia. In contrast, patients with Capgras delusion show intact face perception but believe that closely related persons are impostors. It has been suggested that two deficits are necessary for the delusion, an aberrant perceptual or affective experience that leads to a bizarre belief as well as an impaired ability to evaluate beliefs. Using functional magnetic resonance imaging, we compared neural activity to familiar and unfamiliar faces in a patient with Capgras delusion and an age matched control group. We provide evidence that Capgras delusion is related to dysfunctional activity in the extended face processing system. The patient, who developed the delusion for the partner after a large right prefrontal lesion sparing the ventromedial and medial orbitofrontal cortex, lacked neural activity to the partner's face in left posterior cingulate cortex and left posterior superior temporal sulcus. Further, we found impaired functional connectivity of the latter region with the left superior frontal gyrus and to a lesser extent with the right superior frontal sulcus/middle frontal gyrus. The findings of this case study suggest that the first factor in Capgras delusion may be reduced neural activity in the extended face processing system that deals with inference of mental states while the second factor may be due to a lesion in the right middle frontal gyrus.

* *Corresponding author*. Biological Psychology Lab, Department of Psychology, Carl von Ossietzky Universität, 26111 Oldenburg, Germany.
E-mail address: Christiane.thiel@uni-oldenbur.de (C.M. Thiel).
0010-9452/$ – see front matter © 2013 Elsevier Ltd. All rights reserved.
http://dx.doi.org/10.1016/j.cortex.2013.11.011

1. Introduction

Capgras delusion is a misidentification disorder in which the patient believes that an emotionally closely related person has been replaced by an impostor. The striking feature of Capgras delusion is that patients are able to recognize the related person's face but deny its authenticity. Capgras delusion can occur in schizophrenia and after neurological damage, especially to the right frontal lobe (Edelstyn & Oyebode, 1999). It has been suggested that Capgras delusion represents a mirror image of prosopagnosia, where patients fail to recognize faces but are still able to show affective responses to these faces (Ellis & Young, 1990). The double dissociation thus suggests different neural circuits for face processing, a cognitive one impaired in prosopagnosia and an affective one impaired in Capgras delusion (Breen, Caine, & Coltheart, 2000; Ellis & Young, 1990). While it is known that prosopagnosia is due to damage to right occipito-temporal brain regions, especially the occipital face area (Bouvier & Engel, 2006; Rossion et al., 2003) the dysfunctional brain circuits in Capgras delusion are largely unknown. There are several single case reports using mostly computer tomography (CT) or positron emission tomography (PET) in patients with Capgras delusion which suggest structural and metabolic anomalies in mostly right sided frontal, temporal or parietal brain regions (Horikawa et al., 2006; Lebert et al., 1994; Lewis, 1987; Luca et al., 2013; Paillere-Martinot, Dao-Castellana, Masure, Pillon, & Martinot, 1994). Note that however not all studies found right lateralized structural anomalies (Huang, Liu, & Yang, 1999). Some studies point to the presence of two lesion sites, one in right frontal and the other in right temporal cortex (Alexander, Stuss, & Benson, 1979; Mattioli, Miozzo, & Vignolo, 1999). Functional neuroimaging studies are however missing.

Psychophysiological studies in patients with Capgras delusion provide evidence for impaired autonomic arousal, as measured by skin conductance responses (SCRs), to personally familiar and famous faces (Brighetti, Bonifacci, Borlimi, & Ottaviani, 2007; Ellis, Young, Quayle, & De Pauw, 1997; Hirstein & Ramachandran, 1997). It has thus been suggested that the lack of an autonomic response to an emotionally closely related person may make the patient believe that the person has been replaced by an impostor. The impairment seen in Capgras delusion was proposed to be linked to a disruption of pathways connecting face sensitive areas to limbic cortex, which is involved in the accompanying emotional response (Ellis & Lewis, 2001). Tranel, Damasio, and Damasio (1995) have however shown that patients with bilateral ventromedial prefrontal damage, who also lack autonomic arousal to familiar faces, do not show Capgras delusion, suggesting that the lack of an affective response alone cannot explain the delusion.

Coltheart et al. have suggested a two-factor theory of delusions (Coltheart, 2007; Coltheart, 2010; Coltheart, Langdon, & McKay, 2011; Langdon & Coltheart, 2000). The theory assumes that two deficits are necessary for delusions to occur. The first deficit is an aberrant perceptual or affective experience which will be responsible for the content of the delusion. In the case of Capgras delusion this first deficit may be the lack of autonomic arousal which leads to the abductive inference that the person is an impostor. The second deficit is an impaired ability to evaluate beliefs which prevents the rejection of the bizarre belief. It has been suggested that the anatomical locus of this second deficit is in right frontal cortex.

Functional magnetic resonance imaging (fMRI) studies in healthy volunteers suggest that processing faces of closely related persons involves many brain areas implicated in social cognition, such as the medial prefrontal cortex, the superior temporal sulcus/temporoparietal junction and the posterior cingulate cortex. In most studies activity in these brain regions occurred bilaterally (Gobbini, Leibenluft, Santiago, & Haxby, 2004; Natu & O'Toole, 2011; Shah et al., 2001; Sugiura et al., 2001; Sugiura, Mano, Sasaki, & Sadato, 2011; Taylor et al., 2009). Since these brain regions are even active in implicit tasks and in tasks with newly acquired minimal person knowledge, it has been suggested that neural activity in these areas is related to the spontaneous retrieval of mental states, intentions and attitudes when a familiar face is perceived (Gobbini & Haxby, 2007; Todorov, Gobbini, Evans, & Haxby, 2007). Given the above findings on processing of familiar faces, Gobbini and Haxby (2007) put forward a neural model of familiar face perception that consists of a core system of face processing located in inferior occipital gyrus, fusiform gyrus and posterior superior temporal sulcus that deals with the analysis of visual appearance and an extended system that deals with inference of mental states and emotional responses when seeing familiar faces. In other words, the cores system is responsible for the visual analysis of faces, like early perception of facial features, identification of unique facial identity, as well as eye gaze, facial expressions and lip movements. Faces however convey a wealth of further information, such as emotions, intentions and mental states. This information is extracted by the extended system which involves brain areas implicated in social cognition and emotion such as the medial prefrontal cortex, the superior temporal sulcus/temporoparietal junction, the posterior cingulate cortex and the amygdala. Note that the bilateral posterior cingulate cortex and left posterior superior temporal sulcus seem to be supramodal areas for detecting familiarity since responses were observed for familiar faces and voices (Arnott, Heywood, Kentridge, & Goodale, 2008; Shah et al., 2001). Additionally, the bilateral posterior cingulate cortex was shown to respond to familiar objects and places (Sugiura, Shah, Zilles, & Fink, 2005).

Given this model of familiar face processing it is reasonable to assume that Capgras delusion may also involve dysfunction in the extended face processing system. Hence, according to the Coltheart model, dysfunction in the extended face processing system, leading to a loss of familiarity perception, may represent the first factor leading to the bizarre belief that a loved one is an impostor. Whether brain regions of the extended face processing system are dysfunctional in patients with Capgras delusion when processing familiar faces has however never been tested experimentally by means of functional neuroimaging. A recent single-photon emission computed tomography (SPECT) study in a patient with Alzheimer's disease who developed Capgras delusion provides however first evidence for hypometabolism in left lateralized frontal and posterior midline structures which are parts of the extended face processing system (Jedidi et al., 2013).